Charcot Spine in a Patient With Diabetes Mellitus

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Neuropathic arthropathy is a chronic progressive destructive disorder affecting one or more of the peripheral or axial articulations. The pathophysiology is a disturbance in the normal sensory (pain or proprioceptive) innervation of the joints.[1] The etiology of neuropathic arthropathy has been controversial since Charcot's description in 1868. According to the neurotraumatic theory, which is the most widely accepted, abnormal sensory innervation to a joint will result in rapid joint destruction because of minor traumatic events.[2] Another theory is the neurovascular theory, which assumes that neurologic changes produced by an underlying medical disorder result in a hypervascular region in the subchondral bone that is characterized by increased osteoclastic resorption and osteoporosis. This state leads to pathologic microfractures and eventual subchondral collapse, followed by joint destruction.[3]

Neuropathic spine historically has been related to syphilis.[4] Recently, syringomyelia, diabetes mellitus, congenital insensitivity to pain, and spinal cord injuries have been identified as predisposing factors. The thoracolumbar junction and lumbar spine are the areas most often affected. Neuropathic joints secondary to syringomyelia most commonly affect upper extremities, with the majority being monoarticular, involving the shoulder and less commonly the elbow.[5] There are several case reports of Charcot spinal arthropathy after traumatic paraplegia.[6-12] Our review of the literature showed no reports of Charcot spinal arthropathy secondary to diabetes.

Case Report

Clinical and Imaging Evaluation

A woman in her late 40s, an avid recreational runner, presented with the chief compliant of mild low back and right groin pain. Her problems began 1 year before presentation at our institution, when she complained of an insidious onset of pain in the right groin after a 1.5-hour run. The groin pain resolved, but the low back pain persisted. The patient was evaluated by her family doctor, and lumbar spine radiographs showed marked asymmetrical narrowing of the L1–L2 disc space with some endplate destruction (Figure 1). The patient was initially treated with a nonsteroidal anti-inflammatory drug. There was no improvement, but the pain was mild, and the patient continued to run. Later, computed tomography (CT) and magnetic resonance image (MRI) scans led to a working diagnosis of septic discitis; however, blood work revealed normal white blood cell (WBC) count, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP) level. The patient was then seen by specialists at another institution; needle aspiration of the disc space revealed no organisms. The provisional diagnosis at that stage was still septic discitis, in spite of the negative blood work and disc space aspiration. Four-month repeat MRI scan showed progressive disc space erosion with collapse into kyphosis.

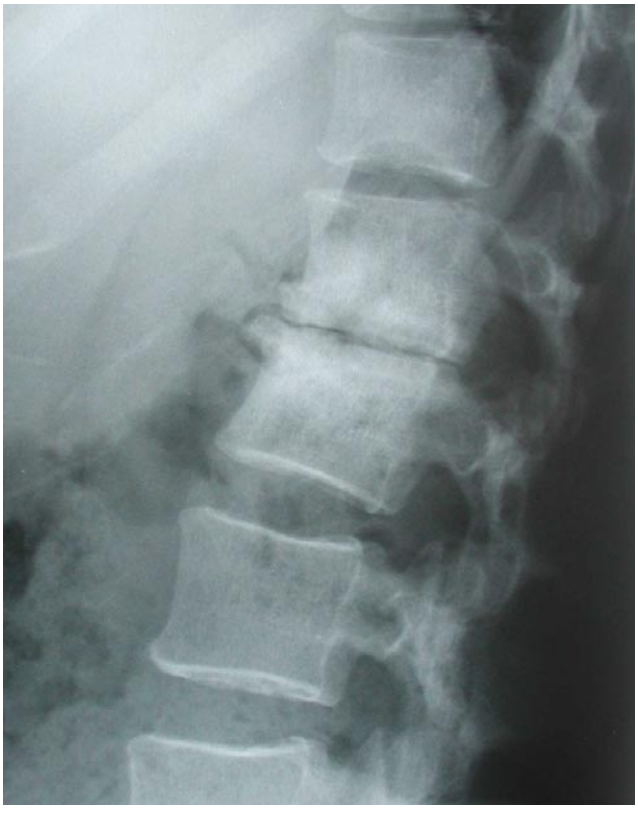

Figure 1. Plain radiograph (lateral view) shows marked narrowing of L1–L2 disc space with some endplate destruction.

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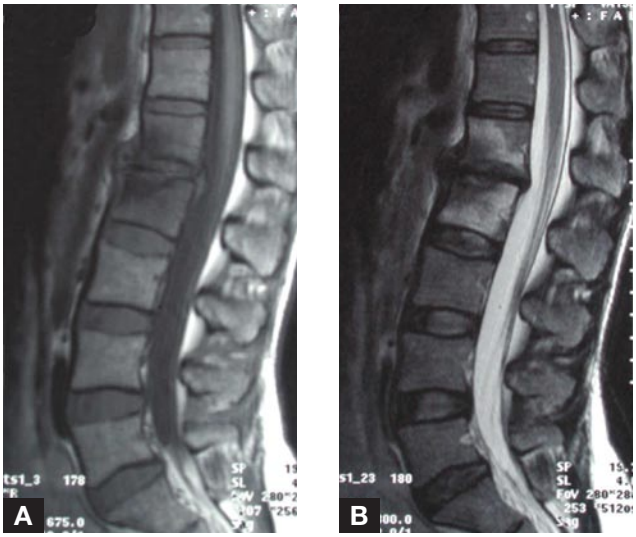

Figure 2. T_1-weighted (A) and T_2-weighted (B) magnetic resonance imaging scans show loss of L1–L2 disc space and endplate erosion with hypointense signal of both disc and endplate, atypical for infection. Hyperintense signal of vertebral body on T_2-weighted scan suggests edema.

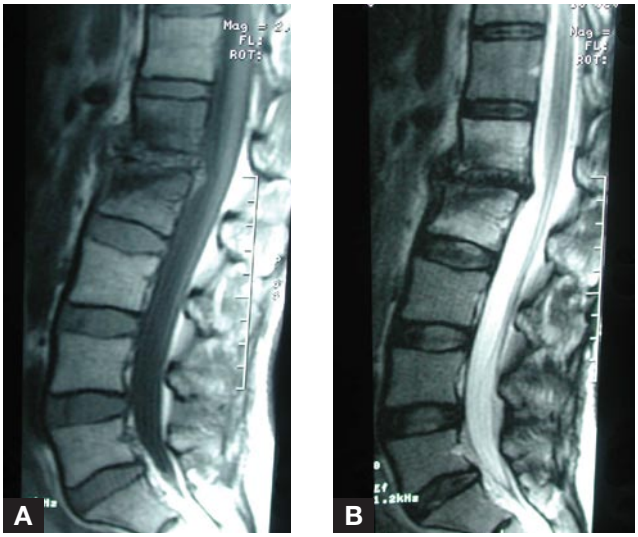

Figure 3. Four-month repeat T_1-weighted (A) and T_2-weighted (B) magnetic resonance imaging scans show progression of disc space changes with some collapse into kyphosis.

Repeat needle aspiration of the disc space was negative. The patient was then referred to our institution, still with the chief complaint of mild low back pain.

The pain was an intermittent ache that ran from the midlumbar region down into the upper buttocks and posterolateral aspect of the hips on both sides. The patient was able to run up to 1.5 hours every day; her back ached only when she ran over a sudden bump. The patient denied a history of fever, chills, or weight loss.

Past medical history included insulin-dependent diabetes mellitus (10 years) and hypertension. The patients was a nonsmoker, had no history of intravenous drug use, and denied any history of trauma or surgery to the spine. Family history and review of systems were unremarkable.

On examination, the spine showed slightly exaggerated thoracolumbar kyphosis. There was no tenderness at the thoracolumbar junction. Range of motion of the lumbar spine was normal. Neurologically, the patient was intact.

The initial MRI scan showed loss of disc space and endplate erosion with decreased signal on T_1- and T_2-weighted images of both the disc and the endplate, atypical for infection (Figure 2). Four-month repeat MRI scan showed progressive disc space changes with some collapse into kyphosis (Figure 3). The rapidity of progression strongly suggested that the underlying pathology was infectious or inflammatory rather than degenerative. The contrast study showed mild enhancement at the L1–L2 level, inconsistent with infection.

The provisional diagnosis was a Charcot neuropathic joint at L1–L2 with differential diagnosis including septic discitis, advanced degenerative disc disease, and tumor. Blood work for infection was negative. Serologic tests for syphilis were negative. Bone scan showed increased activity within the L1–L2 disc space and adjacent endplates. On gallium-67 citrate delayed planar plus tomographic images, low-grade increased activity was present within the L1–L2 disc space. Absence of hyperemia or significant gallium uptake made acute inflammatory process or septic discitis unlikely.

The working diagnosis was Charcot spine secondary to long-standing insulin-dependent diabetes mellitus. The senior author discussed with the patient the risk and benefit of nonoperative versus operative management. The patient decided to undergo operative management in an attempt to prevent further kyphosis, potential instability, and possible neurologic sequelae, as a 4-month repeat MRI scan showed progressive disc space changes with collapse into kyphosis.

Operative Procedure

Surgical management involved T12–L2 instrumentation with transforaminal lumbar interbody fusion (TLIF) at L1–L2. An interbody prosthesis was not used because of concern about inserting a foreign body into the disc space in a patient with suspected osteomyelitis. Autogenous bone was packed into what remained of the extremely narrowed disc space (Figure 4). Intraoperative frozen section and cultures were negative. Intraoperative gram stain showed no bacteria. Histology showed nonspecific bone necrosis without evidence of tumor or granuloma formation. The patient tolerated the surgical procedure well, there were no intraoperative complications, and estimated blood loss was 500 mL. After surgery, the patient was mobilized as tolerated without brace. A prophylactic antibiotic was used for 48 hours after surgery. There were no postoperative complications.

One year after surgery, the patient had local tenderness over the hardware. The fusion appeared solid, as indicated by bridging bony trabeculae across the disc space and

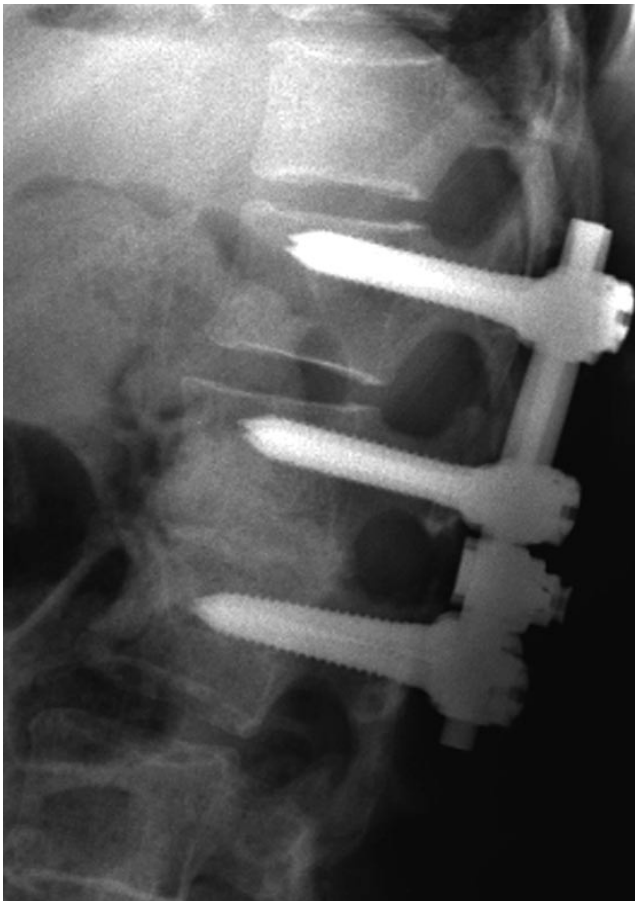

Figure 4. Postoperative plain radiograph (lateral view) shows T12–L2 instrumentation with posterior lumbar interbody fusion at L1–L2. Autogenous bone was packed into remaining disc space.

absence of motion on flexion/extension radiograph. The hardware was removed (Figure 5). By 2-year follow-up, the patient had no pain and had returned to all activities and run a marathon.

The authors have obtained the patient's informed written consent to publish her case report.

Discussion

Spinal neuropathy begins with abnormal movement between 2 or more vertebrae that induces injury to the articular cartilage and subchondral bone. The disc space undergoes degeneration associated with fragmentation of the vertebral endplates, which then progresses to arthropathy, either the atrophic, limited segmental type or the hypertrophic, diffuse type. The atrophic type is less common and shows bone lysis and vertebral body destruction. The more common hypertrophic type is characterized by loss of disc space, bone sclerosis, osteophyte formation, and extensive vertebral ossification. These changes progress to massive periosteal bone formation and bony debris, which lead to a prominent paraspinal mass.[8,12-15] Our patient's initial lumbar spine plain radiograph showed asymmetrical narrowing of the L1–L2 space with more narrowing anteriorly.

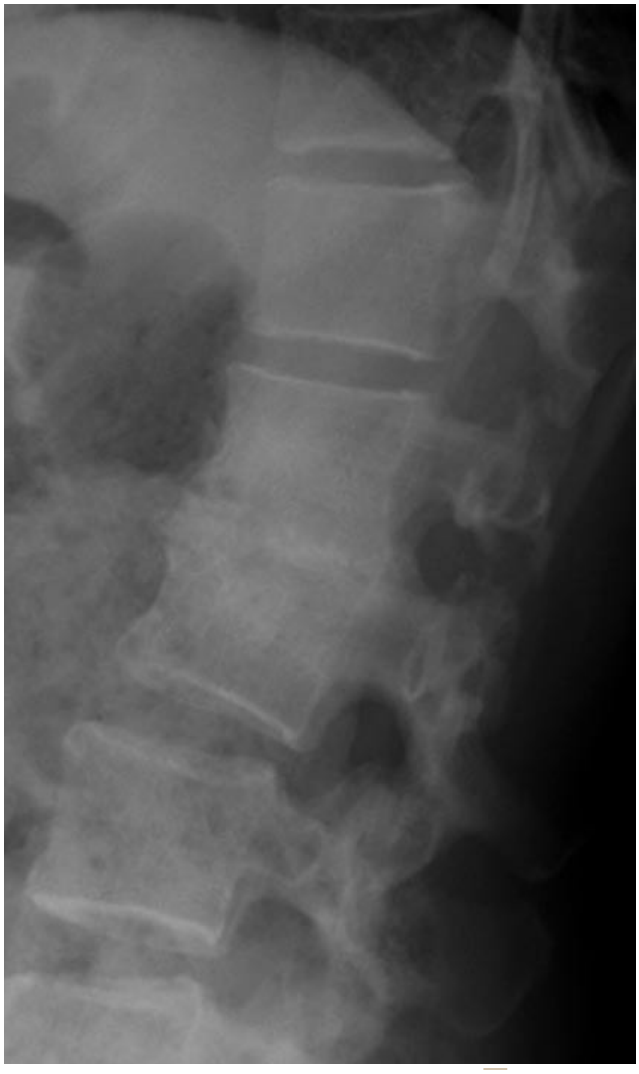

Figure 5. Plain radiograph (lateral view) after hardware removal shows solid fusion.

Although neuropathic joints have been described as painless, patients usually have some degree of pain, though much less than expected based on the amount of joint destruction evident on radiographs.[1,5] Patients usually present with progressive spinal deformity, varying from significant hypermobility to ankylosis. Nerve root compression or bowel and bladder dysfunction can be present in long-standing cases.[16] Our patient presented initially with right hip pain that began after running; the pain progressed to mild thoracolumbar back pain, which did not respond to anti-inflammatory drugs and physiotherapy, though the patient continued to run regularly.

CT provides additional information about the extent of involvement of the osseous structures. Findings include a soft-tissue mass with bone debris replacing the normal disc space, sclerotic and destructive changes in the vertebral bodies and facet joints, an enlarging paraspinal mass with calcifications, and possibly calcification within the spinal canal. These findings closely mimic those in disc space infection with osteomyelitis of adjacent vertebrae.[8,15]

Park and colleagues[15] reported that, on plain radiographs and CT scans, the findings of neuropathic spine were similar to those of disc space infection, severe degenerative disease, and skeletal metastasis. MRI, however, showed characteristic hypointense signal on T_1- and T_2-weighted images, owing to osteosclerotic changes and the presence of surrounding bony debris. Park and colleagues concluded that MRI is helpful in differentiating neuropathy from infection, because on T_2-weighted images the disc space and surrounding bone are of lower signal intensity in neuropathy than in spinal infection. Our patient's initial MRI scan showed loss of disc space and endplate erosion with hypointense signal on T_1- and T_2-weighted images of both the disc and the endplate; these findings were misinterpreted as indicating infection. Four-month repeat MRI scan showed progression disc space changes with some collapse into kyphosis. These findings were not classical for discitis, as there was no fluid within the disc or loculated fluid collection in the region. That our patient's symptoms were relatively mild in light of the imaging changes supported our provisional diagnosis.

The differential diagnosis in our patient's case included severe degenerative arthritis, septic discitis, osteomyelitis, tumor, and Paget disease. WBC count, ESR, and CRP were all normal, which made infection unlikely. Needle aspiration of the disc space, performed twice, was negative for fungus, tuberculosis, and bacteria, as was an open biopsy. Serologic tests for syphilis were negative. Although there is no way to absolutely confirm the diagnosis of Charcot spine, the constellation of clinical, imaging, and laboratory findings strongly supports this diagnosis. The alternative would be an aggressive degenerative disc disease (the location and the rapidity of the aggressive radiographic changes made the probability of this extremely low).

Historically, long-term cast or brace immobilization was used to stabilize the hypermobile segment.[13] Stabilization is difficult given the instability that usually affects the 3 columns and the loss of the painful stimuli that limit activity. Recently, McBride and Greenberg[9] advocated a combined anterior–posterior fusion with instrumentation and bone graft to prevent the disastrous neurologic sequelae that may accompany instability and marked deformity. They used 3-column stability but with a TLIF approach using iliac crest bone graft.

Conclusions

In this report, we emphasize 2 important points. First, Charcot spine is not painless. Patients usually have some pain; the degree, however, is less than expected relative to the amount of joint destruction evident on imaging. Second, Charcot spine should be included in the differential diagnosis of the patient with diabetes and low back pain to prevent unnecessary and potentially risky treatment. Infection, including tuberculosis and fungal infection, should be excluded first, as patients with these conditions are immunocompromised.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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